

#### 242 Consideration on relation between pulmonary status and MBL deficiency in CF patients

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**Background:** Several studies sustain the modulator role of MBL gene on CF pulmonary status. MBL protein is an important factor of the innate immune system that activates complement and opsonisation. Therefore the deficit of MBL can predispose to numerous respiratory infections and deterioration of pulmonary condition. Purpose of study was to determine the relation between patient's pulmonary status and MBL serum levels.

**Methods:** Twenty-three children with age between 6–18 years, DF508 homozygous were tested for MBL protein deficiency. Patients were evaluated by clinical assessment, bacteriological exam and spirometry at last annual evaluation.

**Results:** Among patients with severe pulmonary status (26%) an important part (83% of them) associated low MBL serum levels, while in children with good pulmonary condition only 14.2% were MBL deficient. Relative risk that MBL deficient patient develop severe lung disease is six time superior by comparison to patients with normal MBL.

**Conclusions:** MBL deficiency is a risk factor for severe lung disease in CF children. The increase predisposition for pulmonary infection among MBL deficient patient could be the reason.

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#### 244 Evaluation of an early warning score (EWS) in cystic fibrosis: a retrospective study

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**Background:** In Cystic Fibrosis several independent predictors of morbidity and mortality have been proposed, but the strongest predictor of mortality is considered to be baseline lung function below 30% predicted (Kerem, 1992). In order to identify early those patients at risk from a rapidly declining lung function and clinical status it was decided to evaluate an early warning score (EWS) based on some respiratory independent predictors of morbidity and mortality.

**Objective:** The aim of this study was to evaluate the effectiveness of an early warning score on FEV1 decline and mortality 1 year later.

**Methods:** A retrospective analysis of 140 adult CF patients was conducted. Patients were scored between 0–27 based on baseline and annual decline in lung function, microbiological status, number of IVs/annum, compliance and complications (pneumothorax and haemothorax) at annual review between Jan 2005–2006. The higher the EWS score the greater the clinical concern. The FEV1 decline rate and mortality were assessed one year later.

**Statistical Analysis:** Statistical analysis included Pearson's correlation to examine any relationship between FEV1 decline and EWS and spearman's rank rho for any relationship between number of IV antibiotics and EWS. The Mann Whitney U test to assess any difference between those who died (n=3) and those who survived (n=137) in relation to early warning score.

**Results:** The EWS significantly correlated ( $r=0.59$   $p<0.05$ ) with number of IVs but no other independent variable. Patients who died tended to score higher but this did not reach significance.

**Conclusion:** We were unable to demonstrate any benefit of the EWS over FEV1 on mortality in this study. However, patients with high EWS may benefit from additional MDT input to reduce the number of exacerbations.

#### 243 Sputum investigation in children with cystic fibrosis

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In the last decades numerous scientists pull out sputum investigation as a tool that could be as informative as and more suitable than examination of broncho-alveolar lavage.

We investigated 12 patients with cystic fibrosis (5 girls and 7 boys, aged from 7 to 17 years of age), admitted in the Pediatric clinic for exacerbation more than one time for the last 4 years. The patients expectorated simultaneously and the sputum was collected and studied cytologically and microbiologically. We compared the results from different hospitalisations for each patient.

All the patients were infected with *Pseudomonas aeruginosa*. In 9 patients neutrophil levels are higher in the second admission, which could be explained with the progressive evolution of the disease. In one patient the difference in the neutrophil level between the two hospitalizations is less than 5%. Two of the patients had significant reduction in the neutrophil count. This was probably due to the difference in the *Pseudomonas* strains found out – both of the patients had strain with genes for producing neuroaminidase in the first admission, and strains with no such genes in the second.

Metaplasia in the epithelial cells was found almost in all of the patients, and in 1/3 of them had lower metaplasia percentage in the second admission, which could be connected with the natural history and progression of the disease, but this should be confirmed with further investigations.

**Conclusion:** Early determination of the endobronchial inflammation in CF patients is extremely important for the bacterial infection leads to progressive lung damage long before objective drop in the functional parameters during spirometry test.

#### 245 An audit of the introduction of cough plates in a regional paediatric cystic fibrosis unit

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Isolation of pathogens is central to the appropriate management of respiratory disease in CF. For non expectorating patients; the most common method for obtaining samples has been the cough swab. Cough plates were introduced in to our CF clinics in March 2006 as an alternative and potentially more effective method of sample collection. Cough swabs continued to be used in the ward and community settings.

The aim of this audit was to assess the frequency of positive cultures on cough plates and to determine whether this was comparable to rates previously achieved using cough swabs alone in non expectorating patients aged 3 years and over. Data was audited from March 2005 to February 2006 (Year 1) when cough swabs alone were used and compared to March 2006–February 2007 (Year 2) when both cough swabs and cough plates were in use.

**Results:** 823 cough swabs were taken in Year 1 compared to a total of 782 cough swabs and cough plates in Year 2 (404 cough swabs, 378 cough plates). Positive cultures were obtained in 20% of samples in Year 1 and 18% in Year 2 (cough swabs 20%, cough plates 14%). These are presented in the table. There was no overall decrease in the isolation of pathogens in the year following introduction of cough plates.

**Conclusion:** Introduction of cough plates has not led to any apparent deterioration in standards. However, a large scale study is needed to determine the appropriate use of this method of sample collection and this is currently in progress.

Pathogen	Year 1	Year 2	C swabs (yr 2)	c plates (yr 2)
<i>P. aeruginosa</i>	46	58	30	28
<i>Staph aureus</i>	11	7	4	3
<i>H. influenza</i>	10	17	10	7
<i>Aspergillus</i>	7	4	3	1
<i>B. cepacia</i>	2	0	0	0